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Case report

A rare cause of machinery murmur. Aorta to left lower pulmonary vein fistula: Is it an isolated major aortopulmonary collateral artery?

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Abstract

A female patient, 4 years old having Down syndrome, presented with dyspnea on exertion, repeated chest infections and precordial machinery murmur. Echocardiography, multislice CT scan (MSCT) and angiography defined the presence of an abnormal artery arising from the descending thoracic aorta and ends in the posterior segmental tributary of the left inferior pulmonary vein (LIPV). It resulted in a machinery murmur on auscultation, cardiomegaly and heart failure. A trial of transcatheter occlusion of the arterial fistula failed. The patient was referred for surgery. Left thoracotomy and double ligation and division of the artery were done. The patient had a smooth postoperative course. The machinery murmur disappeared and the antifailure treatment was gradually withdrawn.

Keywords: Machinery murmur; Congenital vascular abnormalities of the lung; Major aortopulmonary collateral artery (MAPCA); Pseudo-sequestration

1. Case description

A female patient aged 4 years having Down syndrome presented to the outpatient clinic in September 2016 with dyspnea on exertion and repeated chest infection. On examination, a continuous machinery murmur was evident in the precordial area and the interscapular region. Having Down syndrome, she had mild physical growth delays, characteristic facial features, and mild intellectual disability. She was maintained on antifailure treatment.

In here medical history, she had an atrial septal defect that was followed by echocardiography till it closed by the age of three years. In early childhood, the repeated chest infections and the cardiac murmur she had was attributed to her ASD. In December 2015, she had an attack of severe pneumonia requiring hospitalization, admission to PICU and mechanical ventilation. The machinery murmur was realized by that time. The echocardiography demonstrated enlarged left atrium and ventricle, normal sized right atrium and ventricle, intact interatrial septum and presence of

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what was diagnosed as major aortopulmonary collateral artery (MAPCA) from descending thoracic aorta to LIPV. The patient's condition started to improve on aggressive respiratory and cardiac medical treatment and she was weaned from mechanical ventilation. Catheterization was decided to visualize the collateral and to obliterate it (Figs. 1–3). The collateral was seen to originate from the descending aorta reaching the LIPV. A balloon was inflated in the artery as a test before coil occlusion for 10 min (Fig. 4). The patient developed sudden decrease of O_2 saturation by 5%. A growing hematoma was also noticed in the groin and the procedure was terminated. The patient was discharged on antifailure measures in January 2016.

The catheterization being done in one of the biggest pediatric cath labs in Egypt, another trial was refused by other centers. MSCTA was done twice for her during the year 2016 confirming the above mentioned vascular data and that

Fig. 1. Aortic injection demonstrated the anomalous artery arising from the aorta and reaching the left inferior pulmonary vein.

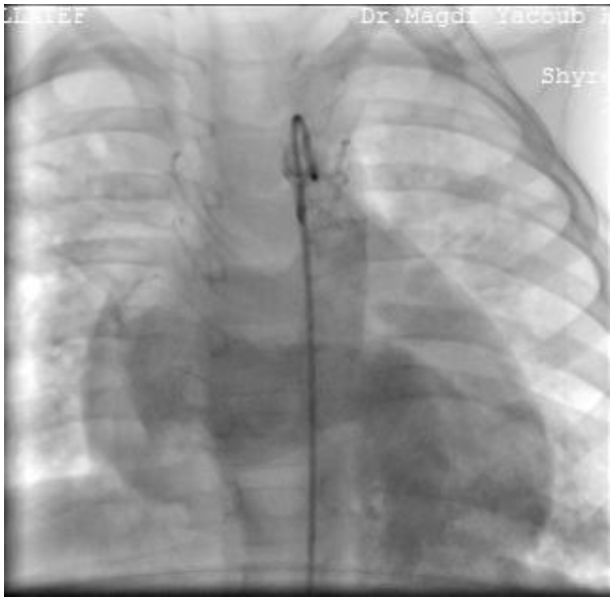

Fig. 2. Rapid left atrial filling after aortic injection of the dye.

Fig. 3. Visualization of left upper and lower lobe pulmonary arteries.

Fig. 4. The patient was reported to develop desaturation after a trial of balloon inflation in the fistulous communication.

the left lung was structurally within normal with no evidence of left pulmonary sequestration. The last CTA done in August 2016 stated the following: Evidence of a sizable collateral artery arising from the anterior aspect of the descending thoracic aorta, averaging 7.5 mm in diameter. It has tortuous course where it curves upwards to connect with the posterior segmental tributary of the left inferior pulmonary vein (LIPV) with resultant dilatation of the LIPV, left atrium and ventricle. The final opinion was anomalous aorta to pulmonary venous fistula with subsequent dilatation of left atrium and ventricle Fig. 5.

The patient was finally referred to our center for surgical closure. High risk consent was taken from the parents to close the collateral with the possibility of doing a left lower lobectomy. Left posterolateral thoracotomy was done. The

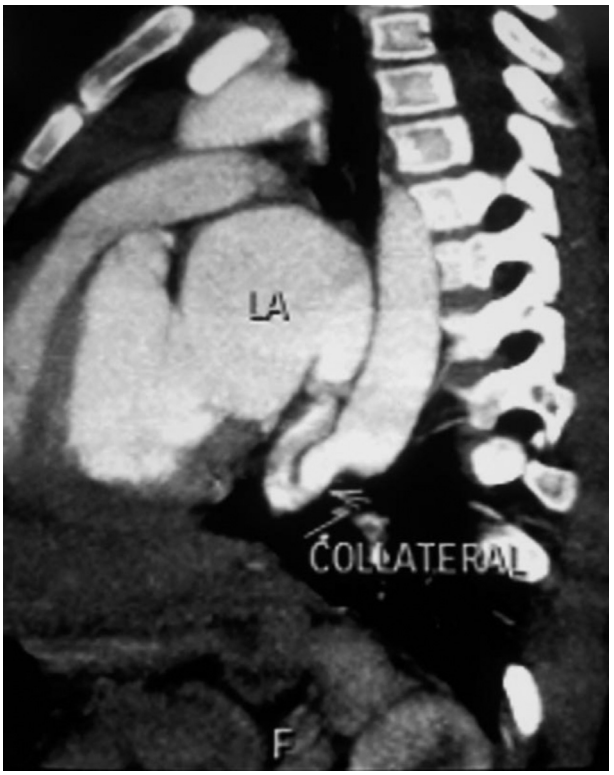

Fig. 5. MSCTA demonstrating the fistulous artery arising from the descending aorta to the LIPV and to the left atrium (LA).

lung was retracted anteriorly. A strong palpable thrill guided us to the site of the collateral. The mediastinal pleura was opened and after careful dissection an artery about 7 mm in diameter, arised from the front of the descending thoracic aorta about 5 cm below the level of the inferior pulmonary vein. In took an S-shaped course to reach that vein. The artery was occluded first for 1 min by forceps and no hemodynamic deterioration or desaturation was observed. It was then doubly ligated at each side and a segment of it was cut Fig. 6. The machinery murmur disappeared. The left lower lobe was completely normal regarding color, texture and inflation and so left lower lobectomy was not done. Hemostasis was done. A single drainage tube was placed and the thoracotomy was closed. The patient was extubated on table with SPO2 100% after extubation. The patient was transferred to PICU in a stable status. In the PICU, the patient passed a smooth post operative course. The patient was discharged from the hospital on the fifth post operative day and was planned for gradual withdrawal of the antifailure treatment.

2. Discussion

Continuous murmur (machinery murmur, Gibson's murmur) is generated by a continuous blood flow shunting from the high-pressure circulation to the low-pressure circulation, throughout systole and diastole [1]. It begins in systole and continues without interruption, encompassing the second sound, throughout diastole or part of thereof [2]. Continuous precordial murmur can be caused by: Patent ductus arteriosus, Coronary fistulae, Palliative surgically created shunts for cyanotic congenital heart diseases, Sinus of Valsalva aneurysm ruptured into right cavities, Aortic-pulmonary window, Atrial septal defect associated with abnormalities that cause increased pressure in the left atrium (Lutembacher syndrome; ASD with mitral stenosis), Left coronary artery origin from pulmonary artery anomaly, Continuous murmur at internal mammary artery ("mammary soufflé" occurs in approximately 10–15% of women at the end of pregnancy and immediately postpartum) and internal mammary arterio-venous fistulae (following implantation of a pacemaker or secondary sternal closure after open cardiac procedures) [1]. Continuous thoracic murmur causes include: Arteriovenous fistula between left supraclavicular artery and left vertebral vein, Pulmonary atresia with MAPCAs and Pulmonary arteriovenous fistula, Truncus arteriosus, Anomalies of origin of the pulmonary artery, Branch of pulmonary

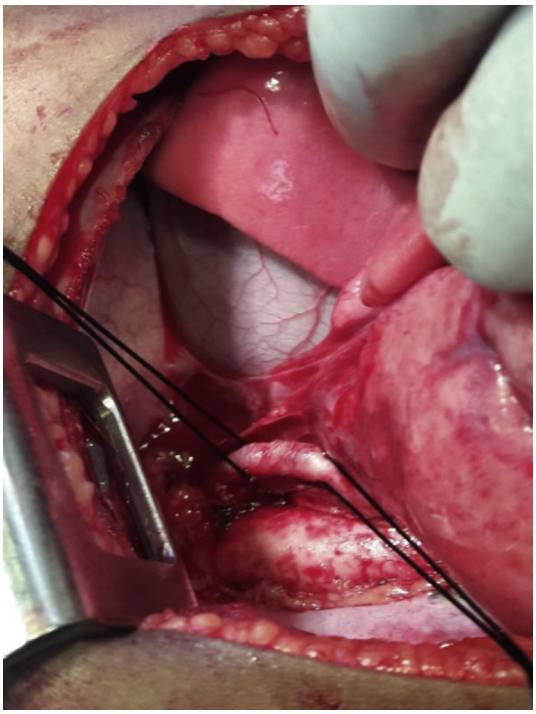

Fig. 6. Through left thoracotomy, the artery was shown to arise from the descending aorta and go to the LIPV and was prepared for ligation and division.

artery stenosis. In cases with coarctation of the aorta, the continuous murmur can be heard only in severe coarctation (narrowing less than 3 mm) in which the developed collateral circulation is not sufficient to ensure normal flow in diastole, thus makes a continuous gradient systolic-diastolic. The murmur is best heard in the posterior chest, inter-scapulovertebral, with maximum of intensity in systole. In some patients with coarctation of the aorta a continuous murmur may be from the dilated intercostals collateral arteries. The anomaly described in this study is an additional reported cause of machinery murmur under this category. Extra-thoracic continuous murmurs include: Venous hum, Cruveilhier-Baumgarten syndrome ("medusa head"), Continuous murmur caused by severe arterial stenosis and Arteriovenous extra-thoracic fistulas (congenital, traumatic or surgically created) [1].

Bronchopulmonary sequestration consists of a nonfunctioning mass of normal lung tissue that lacks normal communication with trachea-bronchial tree, and that receives its arterial blood supply from the systemic circulation. Pseudo-sequestration is mass of normal lung tissue with normal communication with trachea-bronchial tree, and receives its arterial blood supply from the systemic circulation [3]. Two thirds of the time, the sequestration is located in the paravertebral gutter in the posterior segment of the left lower lobe. In this study, although the anomaly was in the lower left side as with sequestration, the left lower lobe connects normally with the trachea-bronchial tree and had its normal pulmonary arterial supply. In addition the artery did not supply any lung tissue but opens directly into the left inferior pulmonary vein. So this anomaly cannot be placed in the spectrum of sequestration or pseudo-sequestration.

Aberrant artery by definition is an artery having an unusual origin or course. A well known example is the aberrant right subclavian artery arising from the proximal descending aorta distal to the left subclavian artery and crosses usually behind the esophagus on its way to the right upper limb. The artery in this study does not apply to the definition of aberrant arteries.

Collateral circulation is the re-routing of blood circulation around a blocked artery or vein via nearby minor vessels. Major aortopulmonary collateral arteries (MAPCA's) are persistent tortuous fetal arteries that arise from the descending aorta and supply blood to pulmonary arteries in the lungs usually at the posterior aspect of hilum. Embryologically, the intersegmental arteries regress with the normal development of pulmonary arteries. They may persist to supply the pulmonary arteries when there is no flow or very little flow into the pulmonary arteries from the right ventricle. Examples of that include pulmonary atresia and severe forms of tetralogy of Fallot. If there is an

alternate supply to the pulmonary arteries (e.g. patent ductus arteriosus), then the fetal arteries regress and such patient does not have MAPCAs [4]. They usually arise from the descending aorta. Less commonly, they may arise from ascending aorta or subclavian arteries. They are usually multiple so that some authors describe them as multiple aortopulmonary collateral arteries rather than major aortopulmonary collateral arteries (MAPCAs). Isolated forms are also reported in the literature [5]. As by definition the MAPCA connect the aorta to pulmonary arteries, the artery described in this study does not fulfill that as it ends in the left inferior pulmonary vein.

Congenital vascular abnormalities of the lung include: Pulmonary artery sling, Pulmonary arteriovenous malformations, pulmonary vein abnormalities, Unilateral absence of a main pulmonary artery, Main pulmonary artery arising from the aorta, Truncus arteriosus, Isolated pulmonary artery aneurysm and Idiopathic hyper lucent lung syndrome (Swyer- James or Macleod's Syndrome). Congenital pulmonary vein abnormalities include: scimitar syndrome, partial anomalous pulmonary venous return, pseudo-scimitar syndrome and pulmonary vein varix.

Congenital pulmonary arteriovenous malformations involve a direct connection between branches of a pulmonary artery and vein. This is also not the case in this study since the artery is the aorta and not a pulmonary artery.

The Latin word fistula literally means tube or pipe. An arteriovenous fistula is an abnormal connection or passageway between an artery and a vein. It may be congenital, surgically created or acquired due to pathologic process, such as trauma or erosion of an arterial aneurysm. Arteriovenous malformation (AVM) is an abnormal connection between arteries and veins, bypassing the capillary system. Both terms can be applied to the studied arterial communication.

From the foregoing, the studied artery can be considered as a separate rare identity among the congenital vascular abnormalities of the lung. It cannot be considered neither in the spectrum of pseudo-sequestrations, aberrant arteries, isolated MAPCAs, nor congenital pulmonary arteriovenous malformations.

Pathophysiologically, as the pressure in the aorta is much higher than in the left inferior pulmonary vein a shunt is created producing the described continuous machinery murmur. A large volume of blood is carried to the left atrium and ventricle resulting in their dilatation and consequently manifestations of ventricular decompensation. Other examination findings may include widened pulse pressures and bounding pulses. In this study the symptoms had taken a progressive course; the artery may had been of small caliber after birth and increased thereafter. This may also explain the delayed discovery of the machinery murmur.

Treatment can be done successfully in the cath lab by coil closure [6,7]. If this failed as what occurred in this study, surgery is indicated to ligate and divide the fistulous communication. As transcatheter coil occlusions do not of course include surgical lobectomies as a part of the procedure to obtain a successful result, in this study left lower lobectomy was not done together with surgical ligation of the fistula with also a good result.

Conflict of interest

No conflict of interest.

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